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CASE REPORT

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Early Prenatal Diagnosis of Thoracopagus Twins by Ultrasound

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ABSTRACT

Introduction: Conjoined twins are identical twins joined in utero. It's a rare phenomenon and present a unique challenge to obstetricians and pediatric surgeons. Conjoined twins are complex complication of monozygotic twinning, which is associated with high perinatal mortality. Case report: At our clinic complete anomaly scan was done, the patients was found to have monozygotic twins of 15 weeks gestation and carrying a conjoint twin. Our ultrasound revealed fully developed heads facing each other, joint at the thorax and sharing a common abdomen. These twins share a single heart with two atrium and two ventricle. They decide for termination of pregnancy after taken the opinion of religious people. Termination of pregnancy was performed by many methods and we chose to use cytotec tablets which inserted vaginally and the outcome was conjoint twin with two bodies fused from the upper thorax to lower belly. Both fetuses are female and died immediately after termination of pregnancy.

Key words: ultrasound, conjoined twins, termination of pregnancy.

1. INTRODUCTION

Conjoined twins are identical twins joined in utero. It's a rare phenomenon and present a unique challenge to obstetricians and pediatric surgeons. The occurrence is estimated to range from 1 in 50,000 births to 1 in 200,000 births, with a somewhat higher incidence in Southwest Asia and Africa (1). Approximately half are stillborn, and a smaller fraction of pairs born alive have abnormalities incompatible with life. The overall survival rate for conjoined twins is approximately 25% (2).

Conjoined twins are complex complication of monozygotic twinning, which is associated with high perinatal mortality. Monoamniotic twins have chances of being conjoined twins if the division of the zygote occurs later than the 12 days, fusion of monozygotic twins is no longer believed to be the basis of conjoined twinning (3). Early prenatal diagnosis of conjoined twins allows better counseling of the parents regarding the management options, including continuation of pregnancy with post-natal surgery, termination of pregnancy or selective fetocide in case of a triplet pregnancy. With the introduction of high-resolution and transvaginal ultrasound, accurate prenatal diagnosis of conjoined twins is possible early in pregnancy (4). However, because of the intrinsic limitation of sonography with regard to tissue contrast, MRI has been explored as a safe alternative (5) as Quad screening (Alpha FP, Beta HCG, S.Estriol, and PAPPA) has a role in diagnosis of N.T.D. Surgery to separate conjoined twins may range from relatively simple to extremely complex, depending on the point of attachment and the internal parts that are shared. Most cases of separation are extremely risky and life-threatening (6).

In this study we present a case of conjoined twin (Thoracopagus) diagnosed prenatally by ultrasound at 15 weeks of gestation and confirmed after termination of pregnancy.

2. CASE REPORT

A 25 years old female patient, primigravida at 15 weeks of gestations, had came for routine antenatal care at our outpatients clinic at Prince Rashid Bin Al-Hassan Military Hospital-Irbid-Jordan. She was seen first by a resident, history and examination was taken, besides routine antenatal investigations then she was referred to our fetal medicine clinic for anomaly scan. There is no family history of twins, and no history of exposure to any medication. At our clinic complete anomaly scan was done, the patients was found to have monozygotic twins of 15 weeks gestation and carrying a conjoint twin. Our ultrasound revealed fully developed heads facing each other, joint at the thorax and sharing a common abdomen. These twins share a single heart with two atrium and two ventricle. A large liver, two stomach, two kidneys and bowel loops were shared by the twin. Also two upper and lower limbs were seen for each fetuses. Both spines were well developed and normal. No other abnormality was seen for two fetuses (Figure 1). So ultrasound diagnosis of conjoined twins of thoracopagus type was made.

The couple was informed about our ultrasound findings and counseled on the various management options. Then the patient went to be seen by another fetomaternal person and

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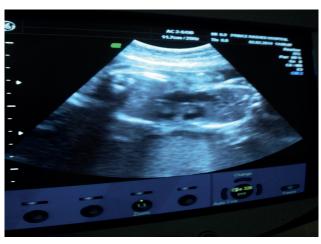


Figure 1. Ultrasound view of thoracopagus twins.



Figure 2. Gross specimens of aborted thoracopagus twins.

they confirmed our diagnosis. They decide for termination of pregnancy after taken the opinion of religious people. Termination of pregnancy was performed by many methods and we chose to use cytotec tablets which inserted vaginally, and the outcome was conjoint twin with two bodies fused from the upper thorax to lower belly

(Figure 2). Both fetuses are female and died immediately after termination of pregnancy.

Diagnosis of conjoined twins was made at 15 weeks, but the decision for termination of pregnancy by parents was taken after 2 weeks, actually at 18 weeks after they taken the opinion of religious people.

3. DISCUSSION

Conjoined twins being the most extreme form of monozygotic twinning, occur in about 1% of monozygotic twins. It is proposed that the origin of conjoined twins is at the primitive streak stage of the embryonic plate (15–17 days), and results from an error in blastogenesis due to incomplete fission of a single zygote. Most conjoined twins are female approximately 70–75%, and most are born prematurely with an extremely high mortality rate (7). Two contradicting theories exist to explain the origins of conjoined twins. The older theory is fission, in which the fertilized egg splits partially. The second and more generally accepted theory is fusion, in which a fertilized egg completely separates, but stem cells (which search for similar cells) find like-stem cells on the other twin and fuse the twins together (8).

Conjoined twins are typically classified by the point at which their bodies are joined. The most common types of conjoined twins are thoraco-omphalopagus (two bodies fused

from the upper chest to the lower chest) which accounts about 28% of cases. The second as in our case is thoracopagus (two bodies fused from the upper thorax to lower belly and the heart is always involved in these cases) which accounts about 18.5% (9).

Antenatal diagnosis by ultrasound is possible in modern day obstetrics. Ultrasonographic identification of any of the following classical signs may suggest the diagnosis: both fetal heads in the same plane, unusual backward flexion of the cervical spine, no change in the relative position after maternal movement and manual manipulations and inability to separate fetal bodies after careful observation (10). Almost all these findings were seen in our case and all feto-maternal person must be familiar with signs associated with conjoint twins and careful ultrasound assessment should be made to identify shared organs.

Early prenatal diagnosis and assessment may provide a window of opportunity to counsel the family for their informed decision on the pregnancy so that early termination can be performed, and to plan for prenatal and perinatal care if pregnancy continued. In report of Wu Y et al, they describe a case of thoracopagus twins diagnosed by fetal echocardiography at 23-week gestation. The 2 hearts fused at the atrial and ventricular levels. They discussed the outcome and review of literature on fetal echocardiographic characteristics of this malformation (11).

Surgical separation of nearly complete conjoined twins may be successful when organs essential for life are not shared. But as in our case they share the same heart, separation of conjoined twins is complicated procedure requiring a multidisciplinary approach and the prognosis is poor as usually predetermined by the underlying anatomy. Consultation with a pediatric surgeon often facilitates parental decision making (12). In our country, there is no cases was reported for separation of conjoint twins, and this help parents for their decision of termination of pregnancy.

Prognosis is very poor among conjoined twins in general. In a study of 14 cases of prenatally diagnosed conjoined twins, 28% of cases died in utero, 54% died immediately after birth, and only 18% survived out of which 50% died postoperatively (13, 14).

4. CONCLUSION

The present case highlights morphological features of an antenataly diagnosed thoracopagus conjoined twins, thus emphasizing the importance of careful antenatal sonographic assessment in all twin pregnancies to rule out conjoined twins. An early diagnosis of conjoined twins is important so that early termination can be performed after counselling couple.

CONFLICT OF INTEREST: NONE DECLARED.

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